Ocular ball bullet injury: detection of gonioscopically unrecognisable cyclodialysis by ultrasound biomicroscopy

EDITOR,—Ocular ball bullet (BB) injuries are vision threatening and more than 1200 people every year are reported to sustain these injuries in the United States. Fewer cases with BB injuries have been reported in Japan. Ultrasound biomicroscopy (UBM) is useful in the morphological evaluation of the anterior segment of the eye. We report a 13 year old boy who sustained cyclodialysis from a BB injury, which was not revealed by gonioscopy but was revealed by UBM. To our knowledge, this is the first report describing detection of cyclodialysis from BB injury by UBM.

CASE REPORT
A 13 year old boy sustained an ocular BB injury in his left eye and visited us the following day. The boy was accidentally shot by his elder brother. The BB was made of plastic. His best corrected visual acuity was right eye, 1.2 and left eye, 0.06. Intraocular pressure was right eye, 17 mm Hg and left eye, 11 mm Hg. Slit lamp examination revealed corneal oedema and Descemet’s folds in his left eye. The anterior chamber was of normal depth and showed moderate inflammation with aqueous cells (1+), flare (1+) and faint fibrinous exudate. Gonioscopy revealed an angle recession inferonasally with a trace hyphaema. The lens, vitreous body, and fundus of the eye were normal. Penetration of the globe was not identified. The patient was treated with atropine 1% three times daily and fluorometholone 0.1% six times daily. The next day, although corneal oedema decreased, the depth of the anterior chamber became shallower than that of the first examination and intraocular pressure decreased to 8 mm Hg. Fundoscopy revealed choriotinal folds in the posterior pole. We performed UBM, which demonstrated a small cyclodialysis in the 5 to 7 o'clock position (Fig 1), which was not apparent gonioscopically. Six days after the injury, the cornea became clearer and no inflammation in the anterior chamber was noted. Even though the depth of the anterior chamber had increased, ciliochoroidal fluid became evident (Fig 2). Intraocular pressure was 7 mm Hg. Ten days after the injury, intraocular pressure increased to 17 mm Hg. Choriotinal folds gradually disappeared and best corrected visual acuity returned to 1.0 six weeks after the incident.

COMMENT
Ocular BB injuries are vision threatening and more than a few of them result in eventual enucleation. However, the patients without open globe injuries have better prognoses. Our patient, who sustained closed globe injury, also regained visual acuity of 1.0. The usual muzzle velocity of a BB gun manufactured in the USA is 350 feet per second and its weight is 0.346 g. Therefore, its kinetic energy is calculated at approximately 2.0 J. In our case, the weight and kinetic energy were 0.2 g and 0.4 J, respectively. We speculate that our patient’s good visual prognosis may be associated with the relatively low kinetic energy generated by the BB gun. Airgun manufacturers’ cooperation in Japan regulates their products to generate kinetic energy of 0.4 J or less. Takashima et al reviewed 50 Japanese patients with ocular BB injury in the literature and described that none of the 50 patients sustained open globe injury and all but one patient had final visual acuity of 0.7 or better. In contrast with the good visual prognosis in Japan, Schein et al reported that 78 of 140 (56%) victims of ocular BB injury in the USA sustained open globe injury and only 31 of 140 (22%) achieved visual acuity of 20/40 or better.

Cyclodialysis is the disinsertion of the ciliary body from the scleral spur and one of its main causes is blunt trauma. Sternberg et al examined eyes enucleated as a result of ocular BB injuries and elucidated frequent damage to the ciliary body histopathologically. The damage included tears into the ciliary body and haemorrhagic necrosis, often accompanied by choroidal haemorrhage and detachment. In the clinical setting, however, it is common that disrupted ocular tissue prevents us from assessing damage to the ciliary body. Additionally, cyclodialysis cleft is often not apparent gonioscopically, even if disruption of the ocular tissue is minimal and the anterior segment is clearly visible. This is because the iris is against the scleral spur, and the cleft is not open as in this particular case. In the series of 140 ocular airgun injuries, the mean age of the victims was 13 years. UBM might be well tolerated by even younger ages because of its non-invasive character. Therefore, the method seems to be useful to evaluate the anterior segment of the patients with closed globe injuries from BB guns.

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Optic canal mucocoele from anterior clinoid pneumatisation

EDITOR,—We describe a 68 year old man who suffered from progressive visual loss in his left eye due to no light perception with afferent and efferent visual field defects, subtle involvement of ipsilateral cranial nerves V(1) and VI. Both computed tomography (CT) and magnetic resonance imaging (MRI) showed a left optic canal lesion with expansion to the superior lateral (anterior clinoid) and inferior walls of the optic canal. Left supraorbital craniotomy was performed by a neurosurgeon. A mucocoele containing turbid fluid in the left strut with compression to the optic nerve was found during the operation. After removal of the lesion, the patient’s best corrective visual acuity was improved to hand movements.

Though visual loss related to mucocoeles of the paranasal sinuses is not rare, the frontal, ethmoid, and sphenoid sinuses are most often implicated. Only rarely has a pneumatised anterior clinoid been reported as a primary location for a mucocoele associated with visual loss. We report a case of anterior clinoid mucocoele producing optic neuropathy and other subtle cranial neuropathies. We emphasise the relation of the optic strut to the optic canal and the superior orbital fissure in producing a symptom complex distinct from optic neuritis and orbital apex syndrome.

CASE REPORT
A 68 year old healthy man noticed progressive loss of vision in the left eye 2 weeks before
admission. He had had frequent attacks of headaches, which were exposed bidigitally via a superior orbital bital craniotomy. Mucoid fluid leaked out as soon as the roof of the optic canal was opened. The postoperative CT scan showed complete removal of the lesion and decompression of the lateral wall of the canal (Fig 2). The patient was discharged 2 weeks after surgery when the visual acuity improved to light perception. In spite of successful decompression, optic atrophy developed eventually. Two years later, after surgery for cataract, the visual acuity in the left eye was no light perception (NLP). There was an amaurotic pupil on the left. There was mild depression of abduction of the left eye. Corneal sensation was mildly impaired in the left eye, and there was no epiphora. No proptosis was present. Ophthalmoscopy revealed normal optic disc, retina, and vessels in both eyes. CT and MRI showed a minimally enhancing lesion of the left anterior clinoid, which encroached upon the superior, lateral, and inferior walls of the optic canal (Fig 1). The optic nerve appeared to be compressed medially. The right anterior clinoid process was pneumatised, suggesting a possible mucocoele of left anterior clinoid process.

Under general anaesthesia, the left optic canal was exposed extradurally via a supraorbital bital craniotomy. Mucoid fluid flowed out as soon as the roof of the optic canal was opened. The postoperative CT scan showed complete removal of the lesion and decompression of the lateral wall of the canal (Fig 2). The patient was discharged 2 weeks after surgery when the visual acuity improved to light perception. In spite of successful decompression, optic atrophy developed eventually. Two years later, after surgery for cataract, the visual acuity in the left eye improved to hand movements.

COMMENT

Mucoceles involving the optic canal are extremely rare. Optic canal mucocele is an ophthalmological emergency, since, without effective management, complete visual loss may develop within a few days. Two reported cases of a mucocele originating in the anterior clinoid process can be found from the literature.1 Both cases demonstrated bilateral pneumatisations of the anterior clinoid. The first patient developed severe visual loss with minimal recovery after surgery. The second patient, who declined surgery, had a recur-rence of symptoms, resulting in optic atrophy. Another case report revealed significant visual improvement from 20/400 to 20/20 after surgery.2 Again, this patient had bilateral pneumatisation of the anterior clinoid, very similar to our presented case. The cause of the mucocele formation is uncertain, since there is no known ostium to become obstructed. Cystic degeneration or secondary inflammation is the proposed mechanism.3 Pneumatisa-


Optic nerve compression by the internal carotid artery in patients with normal tension and high tension glaucoma

EDITOR,—It is generally accepted that ectatic or even normal intracranial blood vessels can cause dysfunction of cranial nerves when situated in an aberrant location.4 Although the occurrence of such compression damage to the optic nerve is poorly recognised, several case reports have shown the optic nerves can in fact be damaged by vascular compression.5 Recently, optic nerve compression by normal intracranial carotid arteries (ICA) has been suggested as a possible cause of visual field defects in patients with optic neuropathy6 and normal tension glaucoma (NTG).7 Various mechanisms have been proposed to explain the aetiology of NTG; however, the exact cause of NTG remains to be elucidated.8

The present study was designed to deter-

10 Three Japanese patients with NTG (average age 65.3 (SD 11.9) years) and 16 age matched patients with high tension glaucoma (HTG) (average 65.6 (12.7) years) were included in this study. Magnetic resonance imaging was performed on a 1.5 T system (Signa Advan-

diate the relationship between the optic nerves and the adjacent ICA, coronal and sagittal T2-weighted images of these structures were taken with the spoilt gradient recalled acquisition in the steady state, one of the magnetic resonance angiography methods. Radiological diagnosis was made by two or three experienced radiologists who were informed of the age and sex but masked to the type of glaucoma of the patients. The relation between the optic nerve and the adjacent ICA was designated as either “with compression” or “without compression” (Fig 1).

In this series, none of the patients had intracranial abnormalities such as tumours, aneurysms, or significant arteriovenous changes of the ICA. In the NTG group, compression by ICA was found in 24 (75%) of 32 optic nerves. Bilateral compression was ob-

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Also had vascular compression. Moreover, there was no statistically significant difference in the clinical characteristics of the eyes with optic nerve compression compared with those without. However, we did find a significantly higher percentage of patients who showed compression of the optic nerves by the ICA in the NTG than in the HTG patients. This difference suggests the possibility that vascular compression by normal ICA may play a role in the visual field defects in some cases of NTG.

**COMMENT**

Compressive optic neuropathy is usually caused by intracranial lesions, such as brain tumours and aneurysms, and not by normal, caused by intracranial lesions, such as brain

**Table 1** Relation between vascular compression and visual function in normal tension glaucoma and high tension glaucoma

<table>
<thead>
<tr>
<th></th>
<th>With compression</th>
<th>Without compression</th>
<th>p Value*</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Normal tension glaucoma</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Number of eyes</td>
<td>24</td>
<td>8</td>
<td>0.786</td>
</tr>
<tr>
<td>Age (years)</td>
<td>65.6 (11.97)</td>
<td>64.3 (11.7)</td>
<td>0.054</td>
</tr>
<tr>
<td>Visual acuity (logMAR)</td>
<td>0.16 (0.44)</td>
<td>0.06 (0.14)</td>
<td>0.083</td>
</tr>
<tr>
<td>Cup to disc ratio</td>
<td>0.80 (0.17)</td>
<td>0.66 (0.23)</td>
<td>0.781</td>
</tr>
<tr>
<td>MD (dB)</td>
<td>-115.02 (7.47)</td>
<td>-15.95 (5.94)</td>
<td>0.698</td>
</tr>
<tr>
<td>PSD (dB)</td>
<td>9.49 (3.70)</td>
<td>11.96 (1.84)</td>
<td>0.082</td>
</tr>
<tr>
<td>CPSD (dB)</td>
<td>8.36 (4.03)</td>
<td>11.39 (3.01)</td>
<td>0.061</td>
</tr>
<tr>
<td><strong>High tension glaucoma</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Number of eyes</td>
<td>12</td>
<td>20</td>
<td>0.321</td>
</tr>
<tr>
<td>Age (years)</td>
<td>68.56 (10.4)</td>
<td>63.9 (13.5)</td>
<td>0.277</td>
</tr>
<tr>
<td>Visual acuity (logMAR)</td>
<td>0.20 (0.27)</td>
<td>0.53 (0.99)</td>
<td>0.757</td>
</tr>
<tr>
<td>Cup to disc ratio</td>
<td>0.62 (0.22)</td>
<td>0.78 (0.23)</td>
<td>0.063</td>
</tr>
<tr>
<td>MD (dB)</td>
<td>-11.23 (10.60)</td>
<td>-15.26 (12.63)</td>
<td>0.361</td>
</tr>
<tr>
<td>PSD (dB)</td>
<td>6.20 (4.27)</td>
<td>5.76 (4.51)</td>
<td>0.795</td>
</tr>
<tr>
<td>CPSD (dB)</td>
<td>5.48 (4.66)</td>
<td>5.01 (4.98)</td>
<td>0.018</td>
</tr>
</tbody>
</table>

*Student’s t test; MD = mean deviation; PSD = pattern standard deviation; CPSD = corrected pattern standard deviation.

**Letters**

Intermittent ptosis due to large exophoria

Editor,—Involuntary closure of the eyelids can be due to either a ptosis with dysfunction of the levator muscle, or a form of blepharospasm with exaggerated contraction of the orbicularis muscle. In true ptosis, no lid crease can be observed in the upper eyelid, whereas in (essential) blepharospasm (secondary) pseudoptosis a lid crease is present.1 We report a case of involuntary intermittent eyelid closure secondary to an exophoria.

**CASE REPORT**

A 67 year old man had complained for 3 years that his left eyelid seemed to fall down and close spontaneously several times a day, and with increasing frequency. This happened especially when he was at home quietly watching television. He was referred to the neurology department of an academic medical centre for treatment of the levator muscle, or a form of blepharospasm with exaggerated contraction of the orbicularis muscle. In true ptosis, no lid crease can be observed in the upper eyelid, whereas in (essential) blepharospasm (secondary) pseudoptosis a lid crease is present.1

When the complaints started 3 years earlier, the patient was referred to the neurology department of an academic medical centre for treatment of the levator muscle, or a form of blepharospasm with exaggerated contraction of the orbicularis muscle. In true ptosis, no lid crease can be observed in the upper eyelid, whereas in (essential) blepharospasm (secondary) pseudoptosis a lid crease is present.1

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The patient was referred to the blepharospasm group of the neurology department of our hospital. There it was noticed that, when the lid opening narrowed, the eye always deviated outward. With the alternate cover test a latent divergent squint was found. Therefore the patient was referred to us for neuro-ophthalmological evaluation.

We found a visual acuity of 20/20 in the right eye and 16/20 in the left, with a hypermetropic correction of about +2.50. Anterior segments, lens, and funduscopy were unremarkable. Pupillary light reactions were normal, and confrontational field testing was full. With close observation in the examination room we saw that the involuntary closure of the left eyelid was always preceded by an exodeviation of the left eye. This was later confirmed with the use of a video camera. After closure, a lid crease could still be observed in the upper lid.

Subsequent orthoptic examination showed a large exophoria (30 prism dioptries) which easily decompenated in a manifest divergent angle (30 prism dioptries). The eye movements were unrestricted and concomitant. The voluntary convergence was excellent. When a manifest deviation occurred, there was mostly suppression of the left eye, although the patient sometimes experienced double vision, especially when asked about it. At reading distance (30 cm), there was some binocular single vision, but not enough to permit examination with a special optometer (TNO stereotest 240°). With the Bagolini striated glasses there was a good fusion area between 20 prism dioptries base temporal and 14 prism dioptries base nasal.

On the basis of these orthoptic results, a presumptive diagnosis was made of a large exophoria of the left eye with a secondary blepharospastic eyelid closure to prevent diplopia. Eye muscle surgery was proposed, and a resection (5 mm) of the lateral rectus and a resection (5 mm) of the medial rectus muscle of the left eye was performed. Postoperatively, the eyes were straight with normal binocular single vision, and no more involuntary eyelid closure or double vision has occurred after a postoperative follow up of 14 months.

COMMENT

In this patient, the easily decompenated exophoria caused diplopia, and this provoked involuntary eyelid closure. The patient was not aware of the diplopia occurring just before his eyelid fell down; he had experienced diplopia occasionally, but could not indicate when. Monocular eye closure in intermittent exotropia has been described and has been thought to be due to avoidance of diplopia. However, Wiggins and von Noorden point out that bright light also may cause monocular eye closure, especially in intermittent exotropes, even when they do not experience diplopia. The authors demonstrated with video recordings that eyelid closure occurred before eleviation in most patients with intermittent exotropia. We did not test our patient under bright light. The patient’s history indicated, however, that the eyelid fell down especially in quiet indoor situations. We also observed it under (dim) examination room lighting. Moreover the patient had good convergence and fusion. Therefore it does not seem to us that bright light played a role in the eyelid closure in this case.

Although in our patient the exophoria had in fact been noticed at several neurological examinations, it had not been recognised as such, or as the possible trigger for secondary ptosis. Neuro-ophthalmological examination at an earlier stage could have prevented unnecessary neuroimaging and botulin injections.

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Permanent extraocular muscle damage following botulinum toxin injection

Permanent extraocular muscle damage following botulinum toxin injection has been reported in a limited number of cases.1–3 While transient side effects like ptosis and diplopia due to local spread of the toxin do occur, to our knowledge permanent extraocular muscle damage has not been reported in a patient with congenital right superior oblique weakness who underwent botulinum toxin injection to the left inferior rectus muscle. This resulted in permanent and profound loss of inferior rectus muscle function, with atrophy of the muscle, confirmed by magnetic resonance imaging scan.

CASE REPORT

A 70 year old white man was seen for increasing angle of deviation of a long standing right hypertropia which had previously been controlled with a small prism correction. His general health was excellent and his only medication was phenelzine 15 mg daily. His visual acuity was 6/5 and distance with right superior oblique underaction, right inferior oblique overaction, left superior rectus underaction, and left inferior rectus overaction. Symptoms of difficulty maintaining binocular vision while reading were initially alleviated by increasing his prism correction, but 9 months later the deviation had increased, measuring 13 dioptries in the reading position and 4 dioptries for distance. A decision was made to proceed with left inferior rectus botulinum toxin injection. The injection was performed under electromyography control using a 27 gauge monopolar needle. The injection was performed through the lower eyelid, angled upwards, advancing the needle posteriorly, superiorly, and nasally by a surgeon (BWF) experienced in the technique. 2.5 U “Botox” botulinum toxin A were injected. The EMG response from the muscle was low to moderate, but there was no apparent complica-

Figure 1 Limited function of left inferior rectus muscle 10 months after botulinum toxin injection

There was a left hypertropia of 20 dioptries in primary position, which increased on laevodepression, in keeping with left inferior rectus muscle paresis. Over the following 10 months, there was persisting diplopia with no change in Hess chart measurements, and no recovery of left inferior rectus muscle function (Fig 1). Forced duction test did not reveal any significant left superior rectus contracture. Investigations for thyroid dysfunction and myasthenia were negative. Magnetic resonance imaging of the patient’s orbits showed atrophy of the left inferior rectus muscle (Fig 2).

Inferior transposition of the medial and lateral recti muscles was performed (by JPL). The inferior rectus muscle insertion appeared normal at the time of surgery. No attempt was made to explore the muscle more posteriorly. The procedure produced satisfactory alignment in primary position, with a small overcorrection in laevoverision.

COMMENT

Injection of botulinum toxin into a clinically overacting muscle produces a temporary reversible paralysis of that muscle. The result of this paralysis is a change in the force dynamics of the paired antagonistic muscles, which allows the weaker opposing muscle to gain force advantage.

The paralytic action of botulinum toxin is attributed to blockade of neuromuscular transmission by interfering with the release of the neurotransmitter acetylcholine at the motor end plate. The paralysis following the use of botulinum is generally associated with complete recovery of neuromuscular function over 3–4 months. Permanent histological changes have been reported in animal studies of the orbital, singly innervated muscle fibre of adult monkey extraocular muscles. Structural changes in muscle fibres and decrease in the density and luminal area of vasculature of the
muscle fibres was seen. The histological evidence of atrophy in the leg muscles has also been reported as a distant manifestation of cervical dystonia. Permanent superior rectus muscle weakness in association with botulinum induced ptosis has also been reported, and was presumed to be due either to breakdown of fusion or contracture of the ipsilateral antagonist. The possible causes of inferior rectus muscle atrophy following botulinum toxin injection in our case include intramuscular haematoma or direct damage to the nerve to the muscle within the muscle cone. Inferior rectus muscle paresis has been reported following retrobulbar anaesthesia for cataract surgery, and the mechanism may have been similar.

Permanent damage to an extrasynaptic muscle following botulinum toxin injection is a rare complication of the procedure. With increasing numbers of patients undergoing the procedure, both the patient and the surgeon should be aware of this rare complication of botulinum toxin injection.

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1 Scott AB. Botulinum injection into extraocular muscles as an alternative to strabismus surgery. Ophthalmology 1980;87:1044–9.


Delayed diagnosis of homocystinuria as a cause of vascular retinal occlusion in young adults

Editor—Retinal vascular occlusions in young adults are seen very infrequently and are generally associated with systemic disorders. Diagnosis of the underlying disease is very important because of treatment and prevention of recurrence. We report two cases initially presenting with systemic neurological disease. Both developed retinal vascular occlusions and the diagnosis of the underlying cause was only made afterwards.

CASE REPORTS
A 30 year old, obese woman presented to the neurologist with acquired perceptive deafness, a tetrapyrinal syndrome, with gait problems and urinary incontinence. Multiple sclerosis was considered, but cerebral magnetic resonance imaging scan showed only atypical lesions. Symptoms gradually decreased without disappearing completely. Six months later, an occlusion of the retinal inferior artery occurred in the right eye, visual acuity was 20/20 in both eyes (Fig 1). Haematological evaluation revealed a disturbed determination of peak levels of homocysteine after oral loading with methionine, suggesting homocystinuria causing thromboembolic processes in the cerebrum and in the eye. Repeated determination after oral loading with methionine after vitamin B12 treatment was still disturbed, confirming the diagnosis of homocystinuria. The patient was treated with pyridoxine and acetylsalicylic acid. Two out of four of the patient’s sisters were also found to suffer from homocystinuria.

A 23 year old obese female, who smoked and had used contraceptives, underwent caesarean section because of negative disagreement and breech presentation. Postoperatively, thrombosis of the arm and erythema nodosum of the legs developed. Four weeks later visual acuity in the left eye deteriorated, caused by massive occlusive retinal vasculitis, retinal haemorrhages, papillitis, and mass central retinal vein occlusion. (Fig 2). A biopsy of the skin lesion of the leg showed leucoclastic vasculitis. Further investigation could not reveal the underlying cause. Treatment with clindamycin, acetylsalicylic acid, and methylprednisolone was started and panretinal laser therapy was performed. Four months after treatment was completed, the other eye developed retinal haemorrhages and occlusion of the upper, temporal branch of the main retinal vein.

Analysis showed elevated peak levels of homocysteine after loading with methionine, suggesting homocystinuria. Analysis of DNA and cystathionine β synthetase in the cultured fibroblasts confirmed the diagnosis of homocystinuria. Treatment with acetylsalicylic acid, folic acid, and pyridoxine resulted in the normalisation of the homocysteine level. The visual acuity 1 year after the first event was 20/15 and 20/50 in the right eye and left eye, respectively.

COMMENT
Homocystinuria consists of a group of different metabolic disorders, all resulting in elevated levels of homocysteine in blood and urine. The most important is the cystathionine β synthetase reduction. Homocysteine interferes with crosslinking of collagen resulting in disturbances in the endothelial synthesis. Also there is an increased platelet adhesion causing thrombotic occlusive disease. The vascular complications led to occlusion of coronary, renal, and cerebral arteries and veins. The frequent manifestations in homocystinuria are disorders of the lens (ectopia lentis, microspherophakia), mental retardation, cardiovascular defects, and skeletal changes. Retinal vascular occlusion is an infrequent manifestation of homocystinuria.

The homocysteine form leads to very early vascular disease with early death and the above mentioned manifestations. Homocystinuric carriers (1:70 in general population) are at risk for occlusive vascular disease at a young age. This can often be prevented by treatment with low doses of acetylsalicylic acid, pyridoxine, and sometimes folic acid. Factors suggesting other causes of thromboembolism (pregnancy, obesity, oral contraceptives) can delay the diagnosis of homocystinuria and subsequent treatment.

In conclusion, homocystinuria should be considered in cases of young adults with retinal vascular occlusions, even if there are no other ocular abnormalities—for example, ectopia lentis. The possible sequelae for general health should be of concern when evaluating and treating these patients.

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Figure 1 Early phase fluorescein angiography of right eye of patient 1, showing occlusion of the retinal nasal inferior artery.
Familial amyloidosis of the Finnish type

EDITOR,—Familial amyloidosis of the Finnish type (FAF), also known as Meretoja syndrome, is a rare autosomal dominant disorder first described by Meretoja in 1969.1 It is thought to develop as a result of a single point mutation involving the gelsolin gene located on chromosome 9. The estimated total number of patients in Finland is 400. Approximately 15 cases have been described outside Finland.2 We present the first case to be recognised in the UK demonstrating the classic signs of corneal lattice dystrophy, cranial neuropathy, and skin changes with an autosomal dominant pedigree.

CASE REPORT

A 73 year old woman presented with gradual reduction in visual acuity in her left eye. She had suffered recurrent corneal erosions affecting her left eye and was diagnosed as having corneal lattice dystrophy 18 years previously. At the time of presentation she was under investigation by a neurologist for progressive weakness of her facial muscles. There was no medical or drug history of note. Family members affected with corneal lattice dystrophy included her daughter and three cousins.

Examination revealed bilateral blepharochalasis, thickened facial skin, and bilateral lower motor neuron facial nerve palsies (Fig 1). Her visual acuity was 6/9 in the right eye and 6/60 improving to 6/36 with a pinhole in the left eye. She had bilateral corneal lattice dystrophy and an area of epithelial loss and sloughing associated with a mild left sided anterior uveitis (Fig 2). Lens opacities were also noted.

She was managed with topical mydriatics, antibiotics, and intensive lubricants. Despite an initial improvement, the cornea failed to re-epithelialise and a combined left penetrating keratoplasty and extracapsular cataract extraction was performed 5 months later. One year postoperatively, her acuity was 6/18 in the right eye, 6/9 in the left eye, and relatively symptom free on intensive lubricants.

The possibility of FAF was considered. Histology of the left corneal button removed at keratoplasty confirmed the characteristic amyloid deposition of lattice dystrophy. Nerve conduction studies demonstrated bilateral facial nerve conduction deficits as well as a subclinical right carpal tunnel syndrome. Scintigraphy using 123 iodine labelled serum amyloid A, and amyloid P. This is in contrast with other cases reported where amyloid stained with antisera to serum amyloid P.3 Whether this represents a subtype of the condition is uncertain.

Various treatments are available targeted at each step in the pathogenesis of all types of amyloidosis with variable success and much research, including genetic manipulation, is being done in this regard.4,5 However, at present the treatment of this disorder is mainly based on alleviating symptoms.

COMMENT

The Finnish type of familial amyloidosis is a systemic disease inherited in an autosomal dominant manner characterised by progressive cranial neuropathy (particularly involving the facial nerve), corneal lattice dystrophy, distal sensorimotor neuropathy, and varying degrees of skin change. The onset of symptoms is typically in the third and fourth decades with slow progress so that the majority are still in good health in their seventh decade.6

The condition is common in Finland but rare elsewhere. This patient and her daughter are the first two cases to be reported in the UK. The patient has the classic features of the disease and demonstrates the point mutation on the gelsolin gene responsible for it. However, although the corneal histology demonstrated the presence of amyloid deposits, immunocytochemistry showed no labelling of the deposits with antibodies to pre-albumin, amyloid A, and amyloid P. This is in contrast with other cases reported where amyloid stained with antisera to serum amyloid P.7 Whether this represents a subtype of the condition is uncertain.

Ocular ball bullet injury: detection of gonioscopically unrecognisable cyclodialysis by ultrasound biomicroscopy

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